Reviews and Bibliographical Notes.

Die Thomsen'sche Krankheit (Myotonia Congenita) Studien. By Prof. Erb, of Heidelberg. F. C. W. Vogel, Leipzig, 1886.

In this monograph of 128 pages Prof. Erb presents as complete an account as can at present be given of Thomsen's disease. It is not merely a compilation of cases heretofore reported, but an entirely independent study of the disease based upon the most careful observations of three cases—two of his own and one sent to Erb by Dr. Fischer, of Canstatt. Having all the methods of examination and investigation thoroughly at his command, Prof. Erb has been enabled to add very materially to our knowledge of Thomsen's disease. The additions he has made are indeed so numerous that a very recent and otherwise able critical digest on this disease in the April number of *Brain* must remain incomplete because it preceded the publication of Erb's monograph by several weeks.

We believe that Erb has now fixed the "type" of this disease, according to which all subsequently reported cases of Thomsen's disease must conform. In view of the general interest of the subject we propose to give a full review of this monograph. We will not pretend to write a *critique;* though we wish to record but one impression, and that is that, to our thinking, Prof. Erb has too rigidly excluded certain variation of symptoms which have been reported by other observers.

In analyzing previous cases the author admits but 23 "typical" cases among those hitherto described. Eleven others he considers doubtful, and among these he classes Eulenburg's case of hypertonia musculorum hypertrophica, about which there has been considerable dispute. He says nothing about two cases referred to by Hamilton, possibly because he has not heard of them, more probably because he does not consider them at all related to this form of disease.

As regards the name of the disease, it will be noticed by the title of the monograph that the author approves of Strümpell's term *myotonia congenita*, and yet retains the name "Thomsen's disease." There is a peculiar propriety in doing this, for the physician who first described it was (is) also a sufferer from the disease.

The chief result of Prof. Erb's studies is the discovery of *typical changes in the mechanical and electrical excitability of the motor nerves and of the muscles ;* and of *marked anatomical changes in the muscles themselves*, which changes are probably closely related to the changes in mechanical and electrical excitability.

The characteristic features of Thomsen's disease are as follows (according to Erb):

The disease is hereditary, and generally affects several members of the same family ; it begins in earliest infancy or as late as the age of puberty.

CLINICAL SYMPTOMS.

Motor Disturbances : After a period of prolonged rest, there is a marked *stiffness* of the muscles and *slowness of movement*, when a movement is attempted, in consequence of which the body may become rigid, the rigidity disappearing only after persistent muscular effort. The phenomena may be exhibited in walking, running, in the attempt to move the hands, and even in chewing. If the patient is asked suddenly to perform a single movement, *e. g.*, to close the hand, he does this promptly enough, but he is not able to relax his muscles at will ; there is evidently a *tonic contraction* outlasting the volitional stimulus. This period of *protracted contraction* may be from 5 to 30 seconds. These symptoms Erb styles *myotonic* disturbances of motion.

The muscles are enormously developed, but their power is not in proportion. In the relaxed condition, the muscles have the normal character ; during contraction they are remarkably hard and firm. Erb observed occasional *fascicular* contractions which he says are distinct from fibrillary contractions,

MECHANICAL AND ELECTRICAL EXCITABILITY.

Motor Nerves : Mechanical excitability possibly diminished, certainly not increased ; *faradic* excitability quantitatively normal ; sudden increase in strength of current may produce prolonged contraction, lasting from 4 to 10 seconds after the withdrawal of the stimulus. *Galvanic* excitability very nearly normal ; single shocks absolutely so ; a succession of shocks may produce tonic prolonged contractions.

Muscles : Mechanical excitability undoubtedly increased ; simple touch with finger being sufficient to excite tonic contraction of the fasciculi under the finger. Such contractions may last from 5 to 30 seconds.

Faradic excitability very marked ; muscles are quickly relaxed when weak currents are used ; sudden increase produces prolonged contraction, but opening contractions, no matter how strong they may be, are never lasting. In one case, keeping the electrode fixed upon a muscle (the gastrocnemius) produced strong oscillating contractions in the muscle. *Galvanic* excitability increased quantitatively (k. c. c. and a. c. c. in different muscles with currents of ¼, 1, 2, m. a.). Qualitative changes : Both poles act nearly equally ; the anode may have a slight preference. The muscular contractions are slow, tonic in character and prolonged. Strong currents invariably produce prolonged contractions. But most characteristic of all are *rhythmical wave-like* contractions produced by stabile currents when the one pole is placed upon the muscle, and best of all, upon its tendinous end. These wave-like contractions proceed from the kathode to the anode, up or down the arm for instance, according to the character of the pole held in the hand. For the electrical phenomena described above, which are typical of Thomsen's disease, Prof. Erb proposes the term *Myotonic Reaction* (My. R.). The R. D. has at last found its mate; the two have some features in common, but in the main they are widely different.

PATHOLOGICO-ANATOMICAL DATA.

From one of his patients Erb had a piece of muscle removed for purposes of examination. In this he found very marked changes, which he considers characteristic of the disease. On cross-section Erb found an enormous hypertrophy of muscular fibres, as compared with the appearance of healthy muscle; the cross-section of each fibre appears well rounded, almost circular, and not polygonal like the cross-section of a normal fibre. The main change, however, is a very considerable increase in the number of nuclei in the sarcolemma; furthermore, an increase of the interstitial connective tissue. In the case of Dr. Fischer, Erb found, in addition to the changes just enumerated, a striking vacuolisation of the individual muscular fibre. The great value of this discovery will be appreciated when we consider that a number of eminent authors were ready to class Thomsen's disease among the heterogeneous functional neuroses.

The question arises, whether or not Thomsen's disease is a disease of the muscles merely. Up to date no changes have been found anywhere else, but the nervous system has as yet been very insufficiently explored. Erb wishes it distinctly understood that he is not willing to affirm or deny that Thomsen's disease may prove to be a tropho-neurosis.

We have given the merest outline of this truly classical monograph. For an account of the differential diagnosis[1] between the clinical symptoms of this disease and those of other somewhat similar forms of disease, and between the pathological changes underlying the various troubles, and for the suggestions regarding treatment, we must refer the student to the original. The monograph is instructive from beginning to end and deserves the very closest study.

We trust that typical cases may soon be reported by American neurologists.

B. S.

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[1] Erb suggests, (p. 79), that hereafter a tap with the percussion hammer and a few closure contractions with the kathode or anode upon certain muscles will be sufficient to establish a diagnosis of the disease. Diseases which may bear some resemblance to Thomsen's disease are: *Tetany, pseudohypertrophy of the muscles, and chronic troubles with marked reaction of degeneration.*